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Cerebral Venous Sinus Thrombosis in Neonates

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Abstract

Neonatal Cerebrovenous sinus thrombosis (CVST) is extremely rare, however it is a devastating condition and one needs to be aware of this condition to diagnose it. The risk factors for CVST are still not properly understood. The largest registry for stroke and for neonatal CVST is from the Canadian registry which quotes an incidence of 0.6 per 100,000 population per year. No data is present for the neonatal CVST in this region.

One needs to be aware of this devastating condition to manage it timely and appropriately. To date there is no consensus on the role of anticoagulant therapy and therefore therapy is largely supportive, however individual cases have to be evaluated and treated on merit.

Introduction

Cerebral venous sinus thrombosis (sinovenous) (CSVT) is an important cause of morbidity and mortality in children. Understanding the natural history of the disease is essential for rational application of new interventions.¹ Venous thromboembolic events in systemic and cerebral vessels are being increasingly recognized in children.

CSVT in children primarily affects neonates and results in severe neurologic impairment and death in some cases. The risk factors for arterial ischaemic stroke and cerebral sinovenous thrombosis in neonates is not well understood. Neonatal arterial ischemic stroke occurs in 1 in 4000 term births. There is a higher risk for boys both in arterial ischaemic stroke and for the sinovenous thrombosis, the reason for the sex predilection is not clear.²

In a recent study of paediatric sinovenous thrombosis in Canada the estimated population incidence was 0.67 per 100,000 children per year. Up till recently the rate of cerebral sinovenous thrombosis in term neonates was unknown. However the minimum incidence of CSVT in neonates in the Canadian paediatric ischaemic stroke reg-

istry was estimated to be 41 per 100,000 newborns per year, this accounts for approximately half of all paediatric cases.³ Cerebro sinovenous thrombosis is however rare in preterm infants.

Presentation

Neonates with CSVT most often present with seizures and lethargy.³ Seizure onset occurs predominantly in the first week of life and very few continue to have seizures beyond the neonatal period.⁴

In Infants with extensive sinovenous occlusion, dilated scalp veins and a bulging fontanelle with splayed cranial sutures may accompany the neurological features.

In older children the presentation is indistinguishable from the benign intracranial hypertension with headache papilloedema and cranial nerve palsies. Cranial neuroimaging done in children presenting with signs of benign intracranial hypertension showed sinovenous thrombosis to be present in 25% of cases.⁵

In the newborn period the brain is still very immature therefore the neurological presentation is limited to only seizures and lethargy. Hemiplegia may occasionally be the presenting sign seen in neonates with CSVT.

Cerebral sinovenous thrombosis is the most frequently recognized cause of symptomatic intraventricular haemorrhage (IVH) in term neonates and is also associated with the presence of thalamic infarcts.⁶ All term infants presenting with intraventricular or thalamic infarcts should therefore undergo further evaluation for the presence of cerebral sinovenous thrombosis.

Following the initial diagnosis there may be propagation of the thrombus without any clinical signs of deterioration. It is therefore very important to be alert to the slightest change in neurological appearance to pick up any extension of the thrombus.

Risk Factors

In childhood CSVT, the thrombosis results from a combination of intravascular and vascular factors. Underlying risk factors including prothrombotic states may predispose to thrombosis, while acute illnesses or prothrombotic medications act as triggers.

The predisposition of neonatal CSVT may relate to physiological factors that may promote CSVT, pathological factors associated with birth process and post natal diseases.

The most frequently involved sinuses in neonate are the superior sagittal sinus and lateral sinuses. Cortical vein thrombosis is rare. The deep sinovenous system involving the Vein of Galen, straight sinus and internal cerebral veins are much less frequently involved

In neonates in addition to the causative factors promoting thrombosis as in older children, the normal moulding and overlapping of the cranial sutures during birth can damage cerebral sinus structures that immediately underlie the sagittal and lateral sinuses promoting CSVT.⁷ In the postnatal period, head positioning alters venous flow within the sinuses.⁸ Perinatal complications reported in neonates with CSVT are usually not obstetrical in nature and are minor.

Asphyxia is a frequent concomitant diagnosis but because the clinical signs of asphyxia are similar to CSVT there is often diagnostic confusion between the two. In a large percentage of conditions, the CSVT may be missed when only asphyxia is diagnosed clinically and no neuroimaging is done.

Prothrombotic abnormalities are also seen in neonates with sinovenous thrombosis although the clear roles of these in the development of sinovenous thrombosis in neonates have not been defined.³

ECMO has been reported to increase the risk of neonatal sino venous thrombosis .Most of the time these neonates do not demonstrate any clinical symptoms as they are paralyzed suggesting that a high index of suspicion is needed for neonates on ECMO. The most likely explanation could be a retrograde thrombosis of the venous system following iatrogenic occlusion of the right of the right internal jugular vein.⁹

A strong association has been reported between preeclampsia , prothrombotic disorders and neonatal venous thrombosis.¹⁰ Dehydration, sepsis and meningitis are common conditions which increase the risk of CSVT in the newborn period.

Although cerebral sinovenous thrombosis has been suggested to play a role in the pathogenesis of IVH (intraventricular haemorrhage) in term neonates the underlying

cause of term IVH is frequently elusive and poorly understood .The possible explanation may be that clot formation in the deep venous structures causes haemorrhage in the ventricles because the deep venous system drains the choroidal, atrial and thalamostriate veins.⁶

Radiographic Features

The diagnosis of CSVT is dependant on either demonstrating decreased flow in the sinuses or the presence of a large thrombus .Non contrast CT scan may demonstrate a large thrombus as increased density and a contrast CT will show a filling defect or a low density area in the concerned sinuses. In children and in neonates computerized axial tomography is a really not ideal as it may miss the diagnosis in at least 10% of cases. The sinuses are adjacent to the bone and may cause interference because of the bony artifact. Occasionally a false positive result may be seen in the non contrast CT because of a high haematocrit, slower blood flow and low density in the adjacent non myelinated brain tissue in the new born brain.¹¹

MRI with MR venogram is the diagnostic tool applicable in early identification of sinovenous thrombosis. CTvenogram with contrast enhancement and with fine cuts taken of the brain may be an alternative if MR is unavailable.

In neonates with an open anterior fontanelle Doppler imaging is also an alternative mode of diagnosis and monitoring of the neonatal sinovenous thrombosis .When the above studies do not clarify the diagnosis, the gold standard would be intravenous digital subtraction angiography.¹²

Recent MRI studies using diffusion weighted imaging have helped in the mechanism of venous thrombosis. The studies have demonstrated early cytotoxic edema preceding the onset of vasogenic oedema. These findings support the presence of primary neuronal injury early in venous infarction. The presence of venous infarction predicts a worse outcome in neonates and children.

The radiographic changes demonstrated in CSVT include parenchymal lesions including venous infarcts which are frequently haemorrhagic and in some newborns transient focal edema may be seen. When the CSVT is extensive, diffuse cerebral swelling with slit like ventricles are seen due to venous outflow obstruction.

Sagittal sinus thrombosis can result in hydrocephalus due to impaired absorption of cerebrospinal fluid into the arachnoid granulations that line the sagittal sinus resulting in increased intra cranial pressure.¹³

Treatment

Anticoagulant therapy for neonatal CSVT is controversial. It is probably not recommended in the presence of a large infarct or significant haemorrhage.

In the Canadian registry of CSVT over one third of newborns with CSVT received anticoagulant medications without major bleeding or extension of the thrombus. Although the potential benefits of the anticoagulant therapy cannot be determined from the registry, the results of the data suggest that haemorrhage was infrequent in the anticoagulated group. However this needs further evaluation especially in the neonates. If anticoagulants are not used radiographic follow-up for subclinical progression is important and if it occurs then anticoagulation could be considered.³

Outcome

Overall neurodevelopmental outcomes have been reported to be good if there is no associated history of asphyxia. Even in the neonates with asphyxia only 50% had neurological sequelae.⁴

The presence of thalamic haemorrhage with CSVT is associated with a poor outcome and cerebral palsy is a common sequelae in the majority of neonates⁵

The overlong term prognosis is really unclear. The best estimate according to the Canadian registry is that after a mean of 2.1 years, 77% of neonates were neurologically normal. However long term follow up is needed since neurological signs are delayed in the neonatal age group.³

Recommendations

In our country the overall incidence of CSVT is unknown. One needs to be aware of this possibility in neonates and look for it in any suspected term neonate with seizures and lethargy. All term neonates with intraventricular and or thalamic haemorrhages should be evaluated for sinovenous thrombosis. The initial investigative tool may be with a Doppler ultrasound and if the suspicion is strong a CT venogram or MRI / MRV should be done.

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